Necrobiotic xanthogranuloma

Necrobiotic xanthogranuloma (NXG) is a rare and chronic skin disorder characterized by the development of yellowish or reddish nodules or plaques on the skin. It is considered a form of non-Langerhans cell histiocytosis, which involves the abnormal proliferation of certain immune cells in the skin.

Here's an overview of key aspects related to necrobiotic xanthogranuloma:

Clinical Presentation:

NXG typically presents as firm, raised nodules or plaques that are usually painless. The lesions often have a characteristic yellowish hue due to the accumulation of lipid-filled cells (xanthoma cells) within the skin. Lesions can occur on various parts of the body, including the face, trunk, and extremities. Histopathology:

Histopathologically, NXG is characterized by the presence of lipid-filled foam cells, inflammatory cells, and areas of necrosis. There may be a chronic inflammatory response around the xanthoma cells. Association with Paraproteinemia:

A notable feature of NXG is its association with paraproteinemia, which refers to the presence of abnormal proteins (paraproteins) in the blood. This is often seen in conditions like multiple myeloma or monoclonal gammopathy of undetermined significance (MGUS). Extracutaneous Involvement:

While NXG primarily affects the skin, it can also involve other organs, such as the eyes (ocular involvement) and other parts of the body (systemic involvement). Ocular involvement can lead to various eye-related symptoms and complications. Treatment:

Treatment of necrobiotic xanthogranuloma can be challenging due to its rarity and variable response to therapies. Therapeutic options may include systemic corticosteroids, immunomodulatory drugs, chemotherapy, and plasmapheresis (a procedure to remove paraproteins from the blood). Other treatments like targeted therapies and phototherapy have been explored in certain cases. Prognosis:

The prognosis of NXG varies depending on the extent of the disease, the presence of associated conditions (such as paraproteinemia), and the response to treatment. Some cases may have a relatively indolent course, while others may be more aggressive and lead to complications. Multidisciplinary Approach:

Due to the potential involvement of multiple organ systems and the association with paraproteinemia, managing NXG often requires a multidisciplinary approach involving dermatologists, hematologists/oncologists, and other specialists as needed. In conclusion, necrobiotic xanthogranuloma is a rare skin disorder with distinct clinical and histopathological features. Its association with paraproteinemia and potential for extracutaneous involvement make it a complex condition to manage. A thorough evaluation by healthcare professionals is essential for accurate diagnosis, appropriate treatment, and ongoing monitoring of patients with NXG.

Popoola et al. report the second observation of an intracranial manifestation and, surprisingly, the first case without the expected dermatologic and systemic dysproteinemia associations. This case identifies an existing knowledge gap in our understanding of necrobiotic xanthogranuloma and

emphasises the need for further research into understanding the presentation, comorbidities and management of this condition [1)]

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Popoola D, Housley SB, Jacoby WT, Lim J, Cappuzzo JM, Levy EI. Non-dermatologic isolated intracranial necrobiotic xanthogranuloma. BMJ Case Rep. 2023 Aug 30;16(8):e254122. doi: 10.1136/bcr-2022-254122. PMID: 37648279.